

**REPORT ON BIRTH DEFECT INVESTIGATIONS CONDUCTED BY THE
TEXAS BIRTH DEFECTS MONITORING DIVISION OR THE HEALTH STUDIES PROGRAM,
TEXAS DEPARTMENT OF HEALTH**

1995

Notes: A glossary of birth defects and other terms is found at the end of this document.

TDH = Texas Department of Health

Because Texas does not yet have a state-wide Birth Defects Registry, expected rates were calculated using data from California, except for the Eagle Pass cluster.

TEMPLE AREA (Bell, Brazos, Falls, Limestone, McClennan Counties)

Condition of Concern: *Gastroschisis.*

Background: A large hospital reported an apparent excess number of gastroschisis cases (7) for the first part of 1994. TDH was requested to assist in an investigation.

Response: TDH conducted a survey of 12 area hospitals in the surrounding counties to determine the number of additional gastroschisis cases. Case medical records were reviewed and state vital statistics analyzed. In total, eight cases were found in 1994. This represents 3.5 times the expected number, and was statistically significant. Further investigation is being planned, and will include interviewing case mothers to get detailed information on potential risk factors.

Duration of Investigation: January - December. Carried over from 1994. 17 person-hours.

GRANBURY (Hood Co.)

Condition of Concern: *Down syndrome.*

Background: TDH received a call from the Texas Department of Mental Health and Mental Retardation of an apparent excess number of Down syndrome cases from Granbury. Seven cases were observed, born from 1992 - 1994. Based on the California rate, 0.49 cases were expected.

Response: TDH conducted an investigation to (a) confirm reported cases and (b) ascertain unreported cases for the town and county. Five cases were confirmed, born to mothers resident in Granbury. This was 10.2 times the expected number, and was statistically significant. Case mothers were interviewed to obtain detailed information on potential risk factors in common that might help to explain the cluster. This

investigation is ongoing, and has been consolidated with the Ellis County investigation.

Duration of Investigation: January - December. Carried over from 1994. 319 person-hours.

MCKINNEY (Collin Co.)

Condition of Concern: *Birth defects related to the ear, specifically atresia of the ear.*

Background: TDH received a call from a citizen in McKinney, of an apparent excess number of children born with ear defects. Six cases were mentioned, born from 1991-October 1994. Based on the rates from California (for any kind of birth defect related to the ear), the expected number of cases in that time period would have been 0.79.

Response: An investigation was conducted to (a) confirm reported cases, (b) ascertain unreported cases for the town. Three unduplicated cases were confirmed (i.e. had been diagnosed by physicians). This was barely statistically significant, the three diagnoses were not the same, and no additional cases have been born in the past two years. No further action required at this time.

Duration of Investigation: January. Carried over from 1994. 15 person-hours.

ELLIS COUNTY

Condition of Concern: *Down syndrome.*

Background: TDH received a call from a concerned citizen about eight cases of Down syndrome, born from 1992 to 1994. This was 1.9 times the expected number based on the California rate.

Response: TDH conducted an investigation to (a) confirm reported cases, and (b) ascertain unreported cases for Ellis county. Four more cases were found from other sources. All 12 cases were confirmed as physician-diagnosed, and born

to mothers residing in the county. This was 2.8 times the expected number, and statistically significant. Available data on the environment were also reviewed. Case mothers were interviewed to obtain detailed information on potential risk factors in common that might help explain the cluster. This investigation is ongoing and has been consolidated with the investigation in Granbury.

Duration of Investigation: January - December. 536 person-hours.

EAGLE PASS (Maverick Co.) and PIEDRAS NEGRAS CITY (Piedras Negras Region, Mexico)

Condition of Concern: *Neural tube defects (NTDs).*

Background: On March 13, the TDH special study Neural Tube Defects Surveillance and Intervention Project team reported an increased number of NTD cases in Eagle Pass and directly across the Rio Grande in Piedras Negras, Mexico. In Eagle Pass during December 1994 to February 1995, two anencephaly cases and one spina bifida case were delivered. These three cases were 13.0 times the expected number of NTDs, based on all 14 counties in the NTD Surveillance and Intervention Project. In the city of Piedras Negras during February and March 1995, three cases with anencephaly (and none with spina bifida) were delivered. Based on NTD Project rates, this was 3.8 times the number of NTDs expected.

Response: Since Maverick County is part of the NTD Project, the three cases have been confirmed. The elevated occurrence (13.0 times expected) was statistically significant. However, there were only two anencephaly cases, and one of the mothers spent some time during her early pregnancy in Mexico. Detailed information on risk factors has been gathered through the case-control study component of the Project. Any additional unreported cases will be identified through the Project's routine active surveillance. TDH personnel met with Mexican health authorities, who confirmed the diagnoses for the reported NTD cases. The ratio of 3.8 times expected was also statistically significant. An offer has been made by the NTD Project to provide the Spanish version of the case-control questionnaire with some biological sampling. TDH also participated in two community meetings with the March of Dimes.

Duration of Investigation: March - present. 170 person-hours.

GAINES COUNTY

Condition of Concern: *Cleft lip and palate.*

Background: On March 31, TDH was contacted

by a citizen concerned about a potential excess of cleft lip and palate. Three cases were reported. Based California's rates, this was 16.11 times the number of cases expected.

Response: TDH is conducting an investigation to (a) confirm reported cases and (b) ascertain unreported cases for the county. The investigation is ongoing.

Duration of Investigation: March - December. 15 person-hours.

DAWSON COUNTY

Condition of Concern: *Spina bifida.*

Background: During the investigation of cleft lip and palate in Gaines County, Early Childhood Intervention staff mentioned concern of high numbers spina bifida cases in the next county to the east, Dawson. Three cases were reported to have been born in 1994. Based California's rates, this was 24.4 times the number of cases expected.

Response: Together with the Gaines investigation, TDH is trying to (a) confirm the reported cases and (b) ascertain unreported cases for the county. The investigation is ongoing.

Duration of Investigation: March - December. 10 person-hours.

THE WOODLANDS (Montgomery Co.)

Condition of Concern: *Anencephaly.*

Background: A concerned couple collected information on potential anencephaly cases in and around The Woodlands for several years. They offered this information to TDH, to assist with investigating a possible cluster in the area.

Response: Using the information, TDH conducted an investigation to (a) confirm the reported cases and (b) ascertain unreported cases for the town. This is ongoing. Some environmental information is also being examined.

Duration of Investigation: August - December. 52 person-hours.

HOUSTON (Harris Co.)

Condition of Concern: *Down syndrome.*

Background: A large hospital in Houston reported a possible excess number of Down syndrome cases delivered there. Six children were born in June and July of 1995. If there had been the same frequency of occurrence as in the prior 10 months, we would have expected only 1.6 cases. (Because this is a hospital-based cluster, it was not compared to expected values derived from population-based registries, such as California.)

Response: Further case finding and case verification were deemed unnecessary in this hospital-based cluster. The six cases were 3.8

times the 1.6 expected cases, based on routine nursery surveillance (i.e., the genetics logs). This was statistically significant. Since this cluster occurred within the pilot study area, Texas Birth Defects Monitoring Division staff will keep close watch on the situation. No further action required at this time.

Duration of Investigation: August - December. 38 person-hours.

DALLAS (Dallas Co.)

Condition of Concern: *Holoprosencephaly*.

Background: TDH received a call from a Dallas

citizen about two cases of holoprosencephaly, a rare birth defect. Born between September and December 1989, the births were unusual in that the mothers lived two blocks from each other at delivery. Between 0.4 and 0.8 cases would have been expected, based on published rates.

Response: The observed number of cases was high, being 2.5 to 4.9 times the expected number. However, with only two cases, it was not statistically significant. No further action is required at this time.

Duration of Investigation: September. 3 person-hours.

GLOSSARY

Anencephaly:	congenital absence of the cranial vault, with cerebral hemispheres completely missing or reduced to small masses attached to the base of the skull.
Atresia:	imperforation; absence or closure of a normal opening.
Cleft lip:	a congenital deformity in which the upper lip is split like that of a hare; also called harelip.
Cleft palate:	a congenital fissure of the roof of the mouth, often associated with cleft lip.
Down syndrome:	a condition characterized by a small, anteroposteriorly flattened skull, short, flat-bridged nose, epicanthal fold, short phalanges, with moderate to severe mental retardation. Due to an extra chromosome 21; one type of trisomy.
Gastroschisis:	a congenital fissure of the abdominal wall, usually accompanied by protrusion of the small and part of the large intestine.
Holoprosencephaly:	impaired midline cleavage of the embryonic forebrain, the most extreme form being cyclopia.
Neural tube defects: (NTDs)	a general term for a number of defects which are presumed to have a common origin in failure of the neural tube to develop properly during the embryonic stage. Conditions include, among others, anencephalus (anencephaly) and spina bifida.
Person-hour:	the equivalent of one person working for one hour; thus 3 person-hours can be derived from 3 people working 1 hour each, or 1 person working 3 hours, or any other combination. This applies only to staff time required in this calendar year.
Spina bifida:	a developmental anomaly characterized by defective closure of the bony encasement of the spinal cord, through which the cord and meninges may or may not protrude.
Trisomies:	having three copies of one chromosome instead of the usual two.

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